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# **Breathlessness and Dysfunctional Breathing in patients with Postural Orthostatic Tachycardia Syndrome (POTS): The Impact of a Physiotherapy Intervention**

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## **Abstract**

Postural orthostatic tachycardia syndrome (POTS) is a chronic, multifactorial syndrome with complex symptoms of orthostatic intolerance. Breathlessness is a prevalent symptom, however little is known about the aetiology. Anecdotal evidence suggests that breathless POTS patients commonly demonstrate dysfunctional breathing/hyperventilation syndrome (DB/HVS). There are, however, no published data regarding DB/HVS in POTS, and whether physiotherapy / breathing retraining may improve patients' breathing pattern and symptoms.

The aim of this study was to explore the potential impact of a physiotherapy intervention involving education and breathing control on DB/HVS in POTS. A retrospective observational cohort study of all patients with POTS referred to respiratory physiotherapy for treatment of DB/HVS over a 20-month period was undertaken.

100 patients (99 female, mean (standard deviation) age 31 (12) years) with a clinical diagnosis of DB/ HV were referred, of which data was available for 66 patients pre – post intervention. Significant improvements in Nijmegen score, respiratory rate and breath hold time (seconds) were observed following treatment. These data provide a testable hypothesis that breathing retraining may provide breathless POTS patients with some symptomatic relief, thus improving their health-related quality of life. The intervention can be easily protocolised to ensure treatment fidelity. Our preliminary findings provide a platform for a subsequent randomised controlled trial of breathing retraining in POTS.

## Introduction

Postural orthostatic tachycardia syndrome (POTS) is a chronic, multifactorial syndrome with complex symptoms of orthostatic intolerance. POTS is defined as an increase in heart rate of  $\geq 30$  bpm in adults (or by  $\geq 40$  bpm in children), within 10 minutes of standing, with no orthostatic fall in blood pressure [1].

POTS is more common in women (5:1), especially those of child bearing age (15-50 years)[2]. Symptoms associated with POTS include: palpitations, light headedness/syncope, chest discomfort, breathlessness, neuropathic pain, chronic fatigue, poor sleep efficiency, gastrointestinal symptoms, syncope, cognitive slowing and psychological distress [3]. The symptom burden for patient with POTS can be high and very debilitating; impacting directly on patient's health related quality of life [4, 5]. The functional impairment in patients with POTS has been compared to that of patients with congestive heart failure and chronic obstructive pulmonary disease (COPD)[6].

Breathlessness is a common symptom in patients with POTS [7, 8]. Cohort studies suggest that  $\geq 65\%$  of patients experience significant breathlessness [9, 10] although little is known about the aetiology in this patient group. Although accurate numbers were not available, anecdotal evidence from our own hospital trust indicates approximately 80 - 90% of patients with POTS have some degree of DB/HVS. Clinically, patients often attribute their breathlessness to changes in breathing pattern, frequency and / or quality of their breathing alongside cardiac symptoms such as tachycardia and / or palpitations. For many patients with POTS, their breathlessness can be episodic, triggered; by physical activity, stressful thoughts and events.

Dysfunctional breathing (DB) is an umbrella term describing breathing disorders in which chronic changes in breathing pattern result in breathlessness and other symptoms in the absence or in excess of the magnitude of physiological respiratory or cardiac disease [11]. There is no consensus definition of dysfunctional breathing, or gold standard diagnostic criteria. The most widely recognised form of dysfunctional breathing is hyperventilation syndrome (HVS), which was first described over 70 years ago [12]. HVS remains the most extensively studied form of dysfunctional breathing. Hyperventilation syndrome is often also

used synonymously with dysfunctional breathing, whereas in fact it is just one type of breathing pattern disorder.

*Boulding and colleagues* [11] suggest the following classification of dysfunctional breathing; **1) Hyperventilation syndrome:** a respiratory pattern associated with symptoms both related to respiratory alkalosis and independent of hypocapnia. **2) Periodic deep sighing:** frequent sighing with an irregular breathing pattern. **3) Thoracic dominant breathing:** a respiratory pattern occurring separate to somatic disease may be considered dysfunctional and results in dyspnoea. **4) Forced abdominal expiration:** inappropriate and excessive use of abdominal muscle contraction to aid expiration. **5) Thoraco-abdominal asynchrony:** a respiratory pattern in which there is delay between rib cage and abdominal contraction resulting in ineffective breathing movements. DB is not a continuously symptomatic state but a syndrome of episodic symptoms that occur with or without recognisable provocation [11, 13, 14].

In clinical practice, patients with POTS often undergo extensive investigations due to the complexity of potential comorbidities e.g. Ehlers - Danlos syndrome (EDS), chronic pain, fibromyalgia and symptoms e.g. chest pain, palpitations, breathlessness, fatigue [30]. There is, however, no guidance or consensus as to which diagnostic test should be undertaken with respect to the assessment of breathlessness in these breathless patients. Anecdotal evidence suggests that breathless POTS patients commonly demonstrate DB/HVS in the absence of structural cardiac and / or respiratory disease [15].

We have, therefore, taken a pragmatic approach to treatment of these patients within our NHS Foundation Trust and they are referred to Respiratory Physiotherapy for assessment and treatment. There are, to date, no published data regarding DB/HVS in POTS, nor the potential benefits that physiotherapy may have on improving patients' breathing pattern and symptoms.

The aim of this paper is, therefore, to describe **1)** the physiotherapy treatment of DB/HVS in POTS, **2)** patients' response to physiotherapy treatment.

## Methods

A retrospective, observational cohort study of all patients with POTS referred to respiratory physiotherapy for treatment of dysfunctional breathing and associated breathlessness between May 2015 and January 2017.

### Respiratory physiotherapy outpatient's referral criteria

Patients with POTS presenting with breathlessness were investigated using a pragmatic approach, within our hospital with the following tests: cardiopulmonary exercise testing, lung function testing, capillary blood gas and a hyperventilation provocation test. A positive hyperventilation provocation test was defined as: end tidal carbon dioxide (etCO<sub>2</sub>) at rest or 10 minutes post voluntary hyperventilation < 4.3kPa.

DB/HVS was identified on the cardiopulmonary exercise test if the following were observed; Hyperventilation was determined by an end-tidal CO<sub>2</sub> (ETCO<sub>2</sub>) <4.0kPa in either the rest, unloaded or pre - anaerobic threshold (AT) parts of the test. Acute hyperventilation was recorded if the respiratory exchange ration (RER) was highly variable or ETCO<sub>2</sub> rapidly normalised at the beginning of the exercise test when unloaded cycling began. Chronic hyperventilation was considered when RER was steady and remained low for a considerable period of the test. Although ventilatory equivalents for O<sub>2</sub> and CO<sub>2</sub> are also often elevated, the definition for hyperventilation is based on the ETCO<sub>2</sub>.

Although difficult to establish, DFB was determined from examination of the ventilatory responses to exercise, with marked variability in minute ventilation (MV), tidal volume (VT) and respiratory rate (RR) considered to be the most robust diagnostic criteria (falls in excess of 5L/min in MV with swings in tidal volume and or respiratory rate). Large swings in the ventilatory equivalents for both CO<sub>2</sub> and O<sub>2</sub> are also observed and reflect the varying efficiency of this breathing pattern.

All patients referred to Respiratory Physiotherapy for assessment and treatment had been extensively reviewed by a Respiratory physician and a Cardiologist and referral based on clinical history, examination, lung function and cardiopulmonary exercise testing. All patients had a positive hyperventilation provocation test, with significant breathlessness either at rest

or disproportionate breathlessness on exertion in the absence of any cardiac and / or respiratory, suggestive of DB/ HVS.

### **Physiotherapy Assessment and Outcome Measures:**

A standardised physiotherapy assessment of patient symptoms was performed on all patients that included: a detailed subjective history including symptom prevalence (see figure 1), review of lung function tests, assessment of chest wall movement / breathing pattern and thoracic posture. Symptom prevalence was quantified by asking patients to give yes / no answer with regards to having any of the symptoms summarised in figure 1.

Assessment of chest wall movement / breathing pattern included: the manual assessment of respiratory motion (MARM), and/ or the Hi Lo breathing assessment [16], respiratory rate and breath hold time [16, 17]. MARM is a palpatory technique, with the patient in the seated position the examiner places their hands over the lower lateral rib cage, where they can assess thoracic expansion and the contribution of the upper thorax and lower thorax/abdomen[17].

The Hi Lo breathing assessment is another palpatory technique. With the patient in the sitting position, the physiotherapist places one hand on the sternum and one on the upper abdomen, to determine whether thoracic or abdominal motion is dominant and to assess for any paradoxical movement. Both MARM and Hi lo are well established clinical physiotherapy assessment techniques used to identify dysfunctional breathing patterns [17].

Patients were asked to complete the Nijmegen questionnaire [18] at each attendance to quantify symptom burden and potential response to treatment.

### **Physiotherapy Intervention**

The physiotherapy intervention was standardised with regards to both the educational component and breathing re-training exercises, which were reinforced using standardised written resources e.g. exercises instruction. A consultant physiotherapist or highly specialised respiratory physiotherapist delivered the intervention. Each patient was assessed and treated

to the point of discharge by the same physiotherapist. The duration of the initial assessment and treatment session was 1 hour, with subsequent sessions lasting 30 minutes.

### **Physiotherapy Treatment**

#### **1. The educational component of the intervention focused on the patient's understanding and consisted of:**

- I. Respiratory physiology, specifically respiratory control and normal breathing pattern
- II. The mind – body link and how external factors such as anxiety, stress and lifestyle influences breathing patterns resulting in breathlessness [19]

#### **2. The breathing re-training intervention**

The breathing re-training method involved teaching breathing control. This approach aimed to achieve nasal breathing, a normal respiratory rate, tidal volume and inspiratory / expiratory ratio with appropriate thoraco-abdominal excursion. The technique is intended to provide patients with a feeling of being in control of their breathing pattern. Breathing control was initially taught with the patient in the supine position. Patients were advised to place one hand on their chest and the other on their abdomen, thereby providing feedback to the patient with regards to reducing any excessive upper chest movements (apical breathing). Patients were then guided through the breathing control exercise which involved breathing with an inspiratory time of two seconds, an expiratory time of two seconds and a post expiratory pause of one second (i.e. total respiratory cycle time = 5 seconds, with the aim of achieving a resting respiratory rate of 12 breaths per minute) [20-22]. If when teaching breathing control, patients were unable to reduce excessive upper chest movements, they were instructed to place both hands behind their head to help fix the upper chest and reduce upper chest motion. Patients were advised that if they were unable to sustain a normal resting respiratory rate (12 bpm) and pattern, or became anxious / experienced significant unpleasant symptoms that they should rest until the symptoms subsided and start again. Patients were instructed that the aim of the breathing exercise was to slowly and progressively increase tolerance of the imposed breathing pattern and rate. All patients were advised to undertake breathing control exercise for 15 minutes twice daily [21, 23].



The breathing retraining exercise programme was standardised and progressive. Once patients were proficient at undertaking the breathing control exercises in the supine position, they were progressed to performing the exercises while sitting and then onto more challenging / functional positions e.g. standing, walking as appropriate.

As patients progressed to the seated posture, the controlled pause technique (exercise) was introduced. The aim of this technique is to desensitise the patient to the ventilatory responses and symptoms associated with the small increases in carbon dioxide that occur when reducing respiratory rate and tidal volume[24]. Following a normal breath in and out patients were instructed to pinch their nose and hold their breath until the first signs of discomfort (air hunger) were experienced. Initially, patients were advised to perform the controlled pause in the supine position, 4 times a day and progressed to the sitting position as symptoms allowed. Once this was achieved, patients were advised to combine controlled pause and breathing control i.e. undertake the controlled pause followed by a sustained period of breathing control [21-23].

All the treatments used in the intervention were standard respiratory physiotherapy techniques commonly applied to patients with acute and chronic respiratory disease [22, 25] their use in the context of breathless POTS patients has not been assessed previously. Patients were discharged from Physiotherapy, when a well-established daily physiotherapy breathing re-training programme had been established and patients were confident to continue the programme independently. Patients that did not attend for follow up appointments were discharged from the service.

## **Data analysis**

All data were tested for normality using Shapiro-wilk normality testing. Demographic data are therefore expressed as mean (SD) as appropriate while Pre – post intervention outcome measures (Nijmegen score, respiratory rate and breath hold time) are expressed as median (range). The significance of change in Nijmegen score, respiratory rate and breath hold time was assessed using non – parametric analysis; Wilcoxon matched pairs signed rank test results summarised in figure2, table 2 All statistical analysis was performed using Prism software (Version 5, GraphPad, San Diego, CA, USA) and statistical significance was accepted as  $p < 0.05$ .

## Results

Over the study period, 100 patients with POTS were referred for Physiotherapy assessment and treatment. All referred patients had a physiotherapy confirmed diagnosis of DB/HVS base on clinical assessment and a Nijmegen score  $>22$  [18]. Patient demographics and characteristics are summarised in table 1. Breathlessness, most notably disproportionate breathlessness on exertion, was highly prevalent in this cohort (figure 1). Of those 100, pre – post physiotherapy data was available for 66 patients. Eighteen patients failed to attend follow up appointments with acute illness, fatigue, burden of hospital attendances and travel distance being the main reasons for non-attendance. There were no marked differences between those patients referred for treatment and completing the intervention (table 1). Sixteen patients were still undergoing treatment at the point that data analysis was undertaken and were therefore excluded from the pre – post physiotherapy data analysis.

Significant improvements were observed pre – post physiotherapy in Nijmegen score, respiratory rate and breath hold time (figure 2, table 2). 97% of patients reported an improvement in symptom burden, quantified by a reduction in their Nijmegen Score, 3% of patient reported a minimal change  $\leq 1$  on the Nijmegen score. 23% of patient reported a Nijmegen score  $\leq 22$ , after receiving the physiotherapy intervention. The mean (range) number of physiotherapy treatment sessions associated with these observed improvements were 3 (2- 8).

**Table 1:** Patient characteristics of all patients with a clinical diagnosis of POTS and DFB/ HV that were 1) referred and 2) completed the physiotherapy intervention. Data are presented as either mean (SD) or number of patients (%)

	All patients referred for Physiotherapy (n=100)	Patients that completed the Physiotherapy intervention (n =66)
Age	31 (12)	32 (13)
Sex	Female = 99, Male = 1	Female = 66, Male = 0
BMI	24 (5)	24 (5)
FEV <sub>1</sub> (L)	3 (0.6)	3 (0.6)
FEV <sub>1</sub> % predicted	98 (13)	96 (13)
VC (L)	4.4 (7)	5(9)
VC % predicted	102 (13)	101 915)
FEV <sub>1</sub> / VC	82 (7)	81 (7)
PEF	416 (70)	410 (67)
PaO <sub>2</sub> (kPa)	12.4 (1.3)	12.2 (1.1)
PaCO <sub>2</sub> (kPa)	4.5 (0.4)	4.4 (0.4)
HCO <sub>3</sub> <sup>-</sup>	24 (1.4)	24 (1.3)
pH	7.46 (0.06)	7.45 (0.03)
SaO <sub>2</sub> (%)	98 (1.4)	98 (1.5)
Smoking status:		
Non-smoker	73 (72%),	52 (79%)
Current	14 (14%)	6 (9%)
Ex-smoker	13 (13%)	8 (12%)
Ehlers–Danlos syndrome (EDS)*	39 (39%)	27 (41%)
Hypermobility syndrome*	19 (19%)	10 (15%)

\* Documented diagnosis as per their clinical notes.

Figure 2: Individual patient changes in (a) Nijmegen score, (b) respiratory rate and (c) breath hold time pre -post physiotherapy intervention. Each line represents an individual patient.

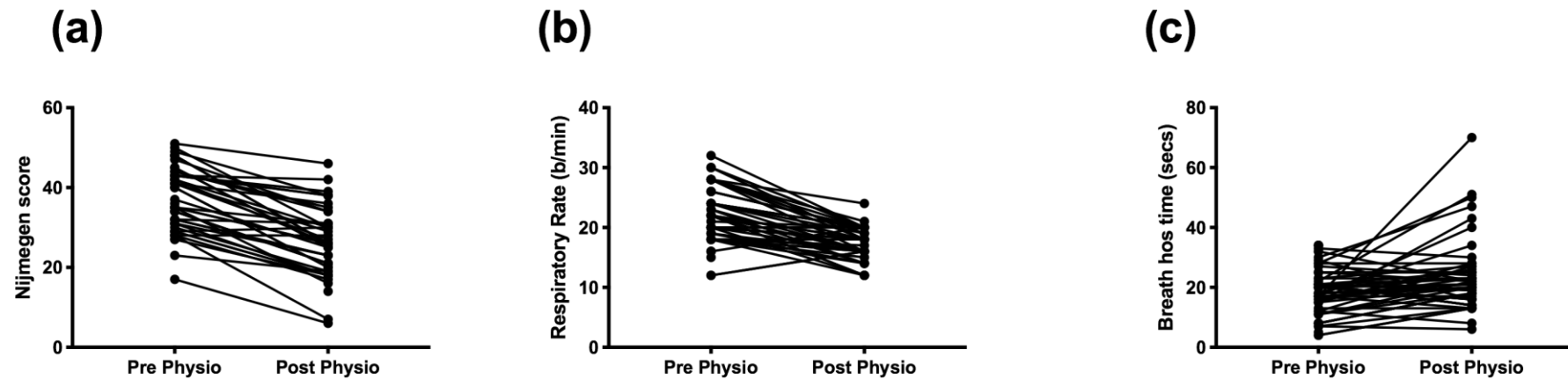


Table 2: Change in outcome measures pre – post physiotherapy intervention. Values reported as median (range).

	<b>Pre-Physiotherapy</b> Median (range)	<b>Post Physiotherapy</b> Median (range)	<b>Median of differences</b>	<b>P Value</b>
Nijmegen score	37 (23 -51)	26 (6-46)	-11	P<0.001
Respiratory rate	22 (12 - 32)	18 (12 -24)	-5	P<0.001
Breath hold time	18 (4-33)	21 (6-70)	+3	P<0.001

## **Discussion**

This paper is the first to characterise dysfunctional breathing/hyperventilation syndrome (DB/HVS) in patients with POTS and to investigate the potential benefit of physiotherapy. The results of this observational case series indicated that a physiotherapy intervention focused on breathing retraining for patients with POTS and dysfunctional breathing, resulted in significant improvements in breathing pattern and symptom burden as quantified by the Nijmegen questionnaire. These data provide a testable hypothesis that breathing retraining may provide breathless POTS patients with some symptomatic relief, thus improving their health related quality of life. The intervention can be easily protocolised to ensure treatment fidelity. The clinical and cost effectiveness of this potential intervention needs to be explored in a subsequent randomised controlled trial (RCT).

### **Significance of the findings**

It is estimated that DB/HVS affects 10% of the general population and is more prevalent in women than in men [26]. Our data demonstrated that patients with POTS who reported significant breathlessness at rest or disproportionate breathlessness on exertion, and / or symptoms of air hunger, excessive yawning or sighing, had DB/HVS. DB/HVS may coexist alongside chronic respiratory diseases such as asthma [26, 27] and chronic obstructive pulmonary disease [28] although whether the relationship is causal or coincidental remains unclear. More frequently DB/HVS occurs in the absence of respiratory disease [13], in keeping with our data.

Although the changes in the Nijmegen score, respiratory rate and breath hold time outcomes for the patient cohort as a whole were all statistically significant post intervention, it is clear from the range of values observed for each outcome measure that not all patients had complete resolution of their DB / HVS following treatment. In addition it is possible that the improvement in breath hold time could have been due a training effect. The lack of an MCID for respiratory rate, breath hold time and Nijmegen limits the clinical interpretation of these observations. Patients were, however, discharged from this service only when they had a

well-established breathing re-training routine and felt confident that they could self-manage their DB/HVS and associated symptoms, supporting the finding that the intervention positively impacted on patient's symptom burden and self-management capabilities. Future studies would undoubtedly benefit from data on MCID of the outcome measures employed and should explore the long-term impact of this type of intervention.

Unexplained breathlessness and '*air hunger*' are the predominant symptoms of DB/HVS[29] and can result in significant patient morbidity and an array of non-respiratory symptoms including: chest tightness, dizziness, palpitations, pins and needles most noted in the feet, hands or around the mouth [22, 29]. The presence of these symptoms can themselves result in anxiety / panic, which can provoke further breathing irregularity.

While orthostatic tachycardia is the hallmark of POTS, orthostasis also initiates increased minute ventilation and decreased end – tidal CO<sub>2</sub> in many patients. Hyperpnea in POTS is related to sympathetic baroreflex stimulation and almost complete cardiovagal baroreflex withdrawal [24] resulting in hypocapnia. POTS patients have decreased central chemosensitivity to CO<sub>2</sub>, but conversely peripheral chemosensitivity to hypoxia is enhanced [30]. It is hypothesised that enhanced hypoxic sensitivity may contribute to the long-term facilitation of sympathoexcitation in patients with POTS, resulting in chronic hyperventilation / dysfunctional breathing patterns. The hypocapnia resulting from chronic / intermittent hyperventilation (HV) reduces cerebral blood flow and contributes to light-headedness and to cognitive impairment during daily life. It also lowers the excitatory threshold for nerve fibres resulting in paraesthesia and reduces the Bohr effect so that haemoglobin releases oxygen less easily. Acute-on-chronic HV results in a reduction in peripheral vascular resistance, and reduced mean arterial blood pressure (MAP). The reduced MAP exacerbates / triggers POTS symptoms of light headedness and fainting, as well as increasing heart rate to compensate for the reduced venous return and lowered cardiac output, and maintain blood pressure [30, 31].

It must be acknowledged that psychological influences such as stress, anxiety and depression, which are highly prevalent in patients with POTS [4, 5] predispose patients to DB/HVS. In our cohort approximately 50% of the patients had anxiety (figure1). Given the high self – reported

prevalence of anxiety and depression in these patients, clinical services and future studies should consider including the GAD-7 anxiety [32] and the PHQ9 – Depression [33]. The impact of our intervention could be, therefore partly related to a change in anxiety achieved by education and greater understanding of DB / HVS in relation to POTS and also empowering patients to take control of their breathing pattern.

Potentially, the observed change in symptoms such as the disproportionate breathlessness on exertion or air hunger, alongside increased breath – hold time may be underpinned by correction of hypocapnia. As hypocapnia is relevant to both the pathophysiology of POTS and the cohort of patients studied. To test this hypothesis future studies should include pre – post PaCO<sub>2</sub> measurements [24].

While it must be acknowledged that it can be difficult in some cases to exclude asthma without undertaking an airway challenge test, in the context of the current preliminary examination of a potential clinical intervention, it was neither feasible nor indicated to undertake provocation testing in all patients. It would be important, however, to precisely phenotype patients in any future large observational multi center studies or randomised controlled trials. Any such diagnostic should include provocation testing, to exclude asthma and because of the presence of unexplained breathlessness a screening echocardiogram to exclude the possibility of pulmonary hypertension.

Our paper suggests that screening for and treating DB/HVS in patients with POTS, reduces symptom burden; the challenge for clinical service provision is that the prevalence of POTS in the UK remains unknown. An interesting observation from our study is that nearly 20% of the patients that were assessed and found to have a DB / HVS failed to attend their follow up appointments. The lack of MCID for the selected the outcomes limited the analysis that could be performed with regards to any exploring factors that might be associated with patients failing to attend their follow up appointment. Given the age of these patients and building on the work of work of *Burton et al*, 2018 [21], a self – guided online breathing retraining intervention may address these issues of nonattendance and equitable access to respiratory physiotherapy.



The provision and access to respiratory physiotherapy outpatient services is limited across the NHS. Many POTS patients are seen by musculoskeletal physiotherapy services due to the coexistence of hypermobility syndromes, chronic pain and chronic fatigue. This paper highlights the importance of increasing awareness of DB/HVS in patients with POTS. This has wider implications beyond respiratory physiotherapy, as clinically the inclusion of a simple breathing retraining approach, alongside other physiotherapeutic interventions may facilitate greater or sustained improvements in physical function and or symptom control in these patients.

The current data highlight the need to determine the prevalence of dysfunctional breathing in patients with POTS, and how this relates to other chronic conditions such as asthma. Alongside this, a core set of clinical investigational standards are also required to enable full phenotyping of patients with POTS and to help understand the factors that influence or predispose these patients to develop dysfunctional breathing patterns. Finally, detailed assessment techniques and classification criteria, which clearly describe the nature of dysfunctional breathing patterns chronic hyperventilation, dysfunctional breathing without hyperventilation or a combination of both, are required. Addressing these unmet needs may help elucidate whether patients with POTS have a propensity for dysfunctional breathing or if such manifestations are a part of the syndrome itself.

### **Limitations**

The data reported are from a service evaluation and hence no control data are available. The improvements observed in patient status could have been due to increased contact with healthcare professionals rather than the treatment intervention. These data should be therefore interpreted with caution. Our findings should be considered preliminary and provide a platform for a subsequent randomised controlled trial.

A limitation of the current outcome measures (Nijmegen questionnaire, respiratory rate and breath hold time) is that they were validated against the hyperventilation provocation test and therefore potentially lack the specificity and sensitivity to detect changes in non – hyperventilation dysfunctional breathing patterns. Future studies may want to consider

techniques, such as structured light plethysmography, that can detect non-hyperventilatory dysfunctional breathing [34].

Due to the observational nature of this study and the small sample size, we were unable to explore the potential role of confounding variables such as pain, deconditioning and anxiety that may affect breathlessness intensity. Applying a randomisation by minimisation approach using baseline Nijmegen score, anxiety score and performance status e.g. EQ5D would help address this issue. When planning future prospective studies, careful consideration will be required to fully characterise the breathless POTS patients enrolled and examine the response of different participant phenotypes to the intervention. Such an approach would facilitate responder analysis. Methodologically, future studies may also benefit from using a randomised, controlled, delayed intervention model, which would address the issue of ensuring an appropriate control group. In addition, a mixed methods approach would allow intervention acceptability, mechanism(s) of action and the impact on patients health related quality of life to be addressed

The patient cohort included in this service evaluation exercise gained access to respiratory physiotherapy due the adoption of a pragmatic approach to treatment. The treatment intervention was based on current evidence in the literature as well as clinical guidelines that advocate breathing re-training for dysfunctional breathing patterns that often co-exist with chronic respiratory disease such as asthma [21, 22]. This may have resulted in selection bias. This was a convenience sample drawn from patients who presented with breathlessness and were subsequently referred to respiratory physiotherapy and then attended. Although limited, the prevalence data, suggests that breathlessness is a significant problem for patients with POTS, supporting the generalisability of these findings to the larger POTS population.

The Nijmegen questionnaire is an established and acceptable outcome measure used within respiratory physiotherapy clinical practice [18, 28, 35, 36]. The Nijmegen score proved useful in characterising these patients, but it must be acknowledged that this tool has not been validated in this patient population. Given the considerable overlapping symptoms with DB / HVS and POTS, the diagnostic utility and sensitivity of the Nijmegen score in POTS needs to be evaluated. Moreover, theses potential limitations of the Nijmegen score highlights the need for a holistic POTS specific patient reported outcome measure that focuses on symptom burden and health related quality of life.

While capillary blood gas and a hyperventilation provocation test the HV screen and EBGs provided a useful physiological outcome measure in the diagnosis of HVS, these are often limited to specialist services and therefore alternative diagnostic tests are sought. Recently, the Breathing Pattern Assessment Tool (BPAT) has demonstrated good sensitivity (0.92) and specificity (0.75) for the diagnosis of dysfunctional breathing in refractory asthma [27]. The BPAT is a novel method of quantifying respiratory pattern / function and may address the challenges of accessing 1) specialist physiological tests i.e. capillary blood gas and a hyperventilation provocation test /or 2) specialist respiratory physiotherapists to provide a clinical diagnosis of DFB/ HV. However, the responsiveness of the BPAT to treatment remains to be ascertained.

Measuring Neural Respiratory Drive (NRD) in breathlessness patients with POTS may be advantageous. NRD provides a global measure of respiratory function and a physiological correlate to breathlessness [37, 38] but, whether NRD is increased in POTS is unknown. To date, there are no data exploring NRD pre – post respiratory interventions such as breathing retraining. Whether these observed improvements in respiratory function (respiratory rate and breath hold) and symptoms are coupled with changes in NRD would be of interest.

## **Conclusion**

In this physiotherapy intervention, which focused on breathing retraining for patients with POTS and dysfunctional breathing, significant improvements in symptoms, respiratory rate and breath hold time were observed following treatment. Although our findings are preliminary, they provide a platform for subsequent investigations and the potential for a randomised controlled trial of breathing retraining in POTS.

## **Ethical approval**

Ethical approval was not required for this study as all patients were receiving routine care provided by King's College Hospital NHS Foundation Trust. Local approvals were obtained prior to undertaking this service evaluation. No funding was provided to undertake this service evaluation.

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